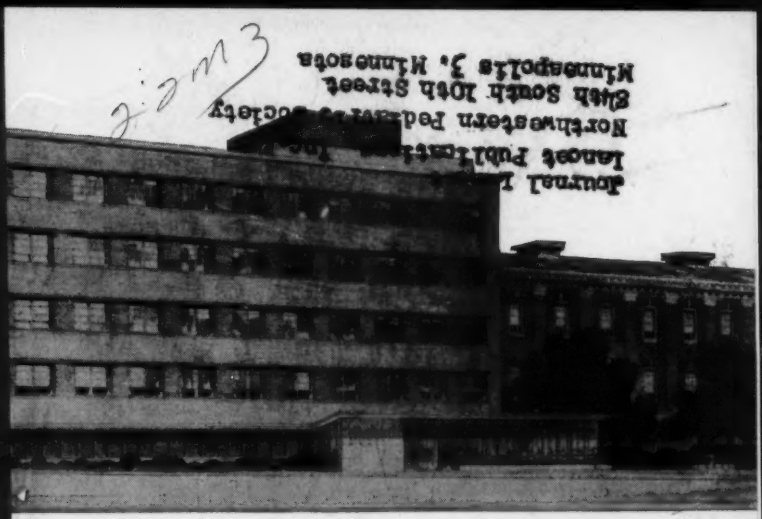

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Clinical Proceedings

OF THE

CHILDREN'S HOSPITAL

WASHINGTON, D. C.

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(B)

VACCINIAL ENCEPHALITIS

Lucas Rubio, M.D.*

It is still not uncommon in the outpatient department of a busy children's hospital to find many pre-school age children who have not been vaccinated against small pox. The potential and serious complication of vaccinial encephalitis due to late vaccination is emphasized with the publication of these two cases occurring during the same week in two children who were vaccinated for the first time at the age of five years.

Case I

This five year old white boy was admitted on September 5, 1955 because of dysphasia of one day duration. Two weeks previously the child had been vaccinated, but there were no untoward effects at that time. Five days prior to admission he had a slight fever in the evening, which persisted until the next day. He also had a slight headache relieved by aspirin. Two days prior to admission he was noted to be quieter than usual, and on the day before admission he awoke complaining of a headache. After breakfast he slept and played indoors. At noontime his temperature was 102.4°F; he refused his lunch, and again slept in the afternoon. He would awaken occasionally, and appeared to behave normally. His family doctor saw him in the evening and prescribed oral oxytetracycline.

On two occasions during the night his temperature was found to be 103° and 104°. During the early morning he was aroused with difficulty and his father noticed that he had a stiff neck, and his speech was unintelligible. The doctor was re-called and hospitalization was advised.

The patient was the product of a normal, full term pregnancy and uncomplicated delivery. Birth weight was 7 pounds, 4 ounces. His growth and development had been normal. He had had measles six weeks prior to admission and chicken pox at the age of 3 years. During the year prior to admission he had fallen twice, striking his head. Skull films taken at both times were reported as normal. Three months prior to admission he again injured his left forehead and had a residual scar as a result.

On admission to the hospital the child was semi-conscious and appeared acutely ill. There was no pallor or cyanosis. A few cervical and submandibular nodes were palpated and the neck veins were distended. A recent vaccination site was noted in the left deltoid region. His pupils were equal, round, and reacted readily to light and accommodation. There was extreme nuchal rigidity and the back was equally stiff. The patellar reflexes were absent; there was a bilateral Babinski sign; there was some hamstring spasm and tenderness on flexion of the thigh. Kernig's and Brudzinski's signs were absent, as were other pathological reflexes. The remainder of the physical examination was non-contributory.

Admission CBC, urinalysis, and serum electrolyte determinations were all within normal limits. Lumbar puncture revealed slightly cloudy pink fluid containing 2,000 red blood cells, and 743 white blood cells per cubic millimeter. Eighty-three percent of the leukocytes were polymorphonuclears. The spinal fluid protein was 60 milligrams per 100 milliliters, and sugar 110 milligrams per 100 milliliters.

During the first eight days of hospitalization the temperature ranged from 99.2° to

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104.6. During the first week of hospitalization the prominent features of the illness were aphasia, flaccid paralysis of all extremities except the right arm, which was spastic, extreme spasm of the muscles of the back and neck and hamstring groups, sluggish pupillary reactions, and a bilateral Babinski sign with absent deep tendon reflexes in the lower extremities. Urinary retention was also reported. The child remained semi-comatose during this entire period.

On the fourth hospital day a second lumbar puncture was performed and slightly hazy spinal fluid was obtained. The protein was 34 milligrams per 100 milliliters and sugar 85 milligrams per 100 milliliters, with 34 leukocytes per cubic millimeter, 96 per cent being lymphocytes.

On the eighth day after hospitalization the child responded to questioning, and there was some return of bladder function, and of muscle tone. All four extremities remained quite weak, but gradual return of muscle power was noted.

Case II

This five year old white boy was admitted to Children's Hospital because he had had a generalized convulsion the day of admission.

Twelve days prior to admission he had been vaccinated for smallpox, this being followed by a local reaction and fever for the following three days. In the afternoon of the day before admission he had had a generalized convulsion which had been preceded by tremors of the face and followed by unconsciousness. The following morning there were more convulsions, which the parents stated began in different places; arms, legs, eyes, and mouth, all progressing to generalized convulsion. He was brought to the out-patient department where he had two more generalized convulsions. These required phenobarbital and light ether anesthesia to control.

Past history, family history, and developmental history were all non-contributory.

On physical examination this was a stuporous white male child who appeared fairly well developed and nourished. There were no significant physical findings except for slight cervical, submandibular and axillary adenopathy. The tonsils were enlarged and somewhat inflamed. Neurological examination was normal except for a marked stupor and a bilateral Babinski sign. There were no other pathologic signs or reflexes.

Initial laboratory examination revealed: hemoglobin 8.9 grams per 100 milliliters; hematocrit 38 per cent; white blood count 6300 with 43 percent polymorphonuclears, 48 per cent lymphocytes, 7 per cent monocytes, and 2 percent eosinophiles. Urinalysis was normal. Spinal fluid contained 92 cells, 54 per cent of which were polymorphonuclears and 46 per cent lymphocytes with 30 milligrams protein per 100 milliliters and 160 milligrams sugar per 100 milliliters.

A few hours after admission the patient was described as appearing "mentally retarded". Over the next few days this mental picture cleared considerably and he seemed much calmer. He was discharged to receive phenobarbital medication and his condition gradually improved.

DISCUSSION

Vaccinial encephalitis will usually occur in children between the ages of four and six years who have been vaccinated for the first time. There is no significant incidence of this complication in children under the age of one year. Although frequently difficult to establish precisely, the period of incubation is usually about 11 days.

The encephalitic signs can appear suddenly or may be preceded by

prodromal signs such as headache or disorientation. The two types are contrasted in the two presented cases. When full-blown, vaccinial encephalitis can present various clinical forms such as encephalitic, encephalomyelitic, myelitic, or meningitic. There is a rather high mortality rate, averaging about 33 per cent. If death does not result, improvement is usually gradual with no sequelae.

In recent publications a familial predisposition has been emphasized. Pre-existing pathology in the central nervous system is also thought to favor this complication. Electroencephalographic evaluation will be of value to discover any subclinical abnormality in this group.

SUMMARY

Two cases of vaccinial encephalitis are presented. This is considered to be a serious illness which is rare before the age of one year and most frequent during the pre-school age period. It is therefore re-emphasized that vaccination should always be performed in children before the age of one year.

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CLINICAL BRIEFS

GANGRENE OF FINGERS AND TOES ACCOMPANYING ROCKY MOUNTAIN SPOTTED FEVER

J. William Oberman, M.D.*

The distinctive gross and microscopic lesions in organs of a patient with Rocky Mountain Spotted Fever are those resulting from thrombosis of small blood vessels. The infecting organism of this disease, *Rickettsia rickettsiae*, lodges in the endothelial and smooth muscle cells of the blood vessel walls giving rise to a proliferative reaction of the endothelium which

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FIG. 1. Early gangrenous changes on the tips of fingers and toes

may soon be followed by thrombosis of the vessel. This pathologic lesion affects arteries, veins, and capillaries and may manifest itself grossly with extensive hemorrhage and necrosis of skin and underlying tissues of the scrotum, ears, face, fingers, toes, and vulva.

Clinical Data

An engorged tick was removed from the cheek of this 16 month old white girl two weeks prior to hospitalization. One week prior to admission, high fever and delirium developed. Two days later a rash appeared, initially being maculopapular in type and rapidly becoming purpuric, and distributed over the entire body but more marked on the extremities. High fever of 104–106° continued and was accompanied by signs of extreme toxicity. On the ninth day of illness a bluish discoloration of the fingers, toes, and tip of the nose was noted.

Agglutination titer against *B. proteus* strain OX19 rose from 1:160 during the first week of illness to 1:320 during the second week. Therapy with chlortetracycline and chloramphenicol was instituted and although the clinical course was stormy the patient gradually became afebrile on the twentieth day of illness. Recovery was complete except for the presence of dry gangrene of the terminal phalanges of the fourth and fifth fingers of the right hand and the four small toes of the right foot.

BULLOUS LESIONS IN THE NEWBORN INFANT

Julia Adler, M.D.*

The differential diagnosis of rashes or skin lesions in infancy is sometimes quite difficult because they often lack any typical characteristics. This is more true of younger than older infants. In the newborn period, for example, one speaks of seborrheic, eczematoid, bullous, or pemphigoid lesions without further differentiation. The exact differential diagnosis is, of course, quite important, because: 1) some of the skin diseases are contagious, 2) the prognosis varies widely, and 3) the treatment will usually be different.

When bullous lesions appear on the skin of a newborn infant, the following entities should be considered:

1. On rare occasions congenital *bullae* have occurred on the skin of an otherwise normal infant. During delivery these bullae may or may not rupture so that the skin of the infant may demonstrate bullae containing clear fluid and in addition large denuded areas of skin. These bullae have been reported to appear between the third and eighth month of pregnancy. Initially their size is 0.5–1.0 centimeter in diameter but subsequent lesions are sometimes much larger. Following rupture and in the absence of secondary infection, these lesions dry up in a few days eventually leaving normal skin. The course is benign and the general health of the infant remains good.

2. *Pemphigus neonatorum* is a contagious skin disease of early infancy characterized by the appearance of large blisters scattered over the trunk and extremities which develop between the fourth and seventh day of life. The palms and soles are not involved. These large vesicles are of varying size, usually being less than 1 centimeter in diameter. There is no surrounding redness or edema. They contain serous fluid from which staphylococci or streptococci may be cultured. With rupture of a bullus, new skin areas become infected. The general condition of the infant is usually good and there is usually no fever; the appetite is unimpaired; and early development is normal.

The disease is usually contracted from individuals with impetigo, furuncles or abscesses who handle these young infants. Since it is highly contagious, each patient must be isolated. Antibiotic or sulfonamide therapy will usually result in a complete cure; the prognosis in general is good. Individual lesions usually heal in one to two weeks without scarring.

3. *Ritter's disease*, which was originally described in 1878 under the name dermatitis exfoliativa neonatorum, as a rule appears during the

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first or second week of life. The initial manifestation is usually perioral redness and edema which later spreads over the entire trunk and extremities. Characteristic exfoliation follows (second stage) and the skin peels off in large plaques.

Ritter's disease may also occur in another form, the onset of which is characterized by the appearance of blisters of varying size which enlarge rapidly and eventually involve the entire skin. Many of these bullae rupture, leaving behind denuded areas which resemble severe burns. Areas of apparently normal skin may be peeled off very easily by mild friction (Nikolsky sign).

Prognosis in spite of the early and adequate use of antibiotics is poor. The general condition of these infants deteriorates rapidly: they have no appetite; often have fever; and are frequently subject to complicating diarrhea or pneumonia. Sepsis is common and is frequently fatal. Finkelstein (1920) reported a mortality rate of 60 to 70 per cent. With modern therapy, mortality is probably still 45 per cent.

The etiology of Ritter's disease is unknown. Many authors believe that the disease is a malignant form of pemphigus and do not acknowledge it as a separate disease. In addition to the difference in prognosis there are other well defined differences between the two diseases. Pemphigus neonatorum almost always starts on the trunk while Ritter's disease begins on the face, later spreading to the trunk and extremities. In the course of pemphigus, many exacerbations and remissions occur whereas in Ritter's disease, only one eruption occurs. Finally, pemphigus neonatorum lacks the typical exfoliation which characterizes Ritter's disease.

Microscopic examination of the skin in Ritter's disease reveals inflammatory changes in the corium with infiltration by leukocytes, histiocytes and a few lymphocytes. There is separation of the epidermis from the underlying tissue. The deeper layers of the skin are not involved.

4. The bullous lesions of *syphilis* do not usually give rise to any difficulty in differential diagnosis due to the characteristic localization of the lesions, other signs of congenital syphilis, a positive serological test for syphilis, and a history of maternal syphilis, either inadequately treated or entirely untreated. When large doses of penicillin are given the prognosis is good.

5. *Acrodermatitis enteropathica* usually develops later than the previously described diseases. The lesions occur as irregularly shaped vesiculopustulobullous plaques located around the mucocutaneous orifices and on the distal portions of the extremities. There is frequently an accompanying alopecia; paronychia and distorted nails are common; and photophobia is very often present. The disease is characterized by periods of exacerbation and remission. With exacerbations these infants have diarrhea and may have symptoms of the celiac syndrome. The prognosis is variable

but in some cases the disease may be controlled by medication with Didoquin®.

6. *Epidermolysis bullosa* is well known as a benign hereditary skin disease. It was originally described by Kobner in 1886. The disease is inherited dominantly and usually has its onset in early childhood or even later. Bullae may appear in any location although the sites of predilection are areas exposed to mechanical irritation such as the neck, elbows, fingers, etc. General health is good and prognosis for life is excellent.

Hallopeau a few years later described the dystrophic form of epidermolysis bullosa which he termed "forme bulleuse and atrophic". Inheritance of this form is variable. The skin lesions heal with atrophy and scarring. The prognosis for life is good.

The first reported cases of epidermolysis bullosa congenita were described by Jenny, Linser, and Ritschel (1920–1924). They reported four infants born of healthy parents who married consanguineously. There was no history of familial disease, and pregnancy and delivery were normal. These infants showed epidermal defects or bullae at birth or in the first two days of life. These lesions became widespread in a very few days. The general condition of these infants deteriorated rapidly and death occurred within six to fourteen days after birth.

Herlitz, in 1935, summarized several cases in newborn infants of a condition which he called epidermolysis bullosa congenita hereditaria lethalis. This is a very rare and invariably fatal type but is not necessarily inherited. Death usually occurs in four to six weeks in spite of all treatment.

Langhof in 1952 claimed to be able to prevent the appearance of new bullae in epidermolysis bullosa hereditaria lethalis with an ointment containing heparin. ACTH and cortisone are of no value in the therapy of this disease.

SUMMARY

The differential diagnosis of bullous lesions occurring in the newborn period is described. The prognosis and therapy of each condition is compared and contrasted.

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RHEUMATIC FEVER—I

(Lecture Review)*

Bernice Wedum, M.D.†

Edited by J. William Oberman, M.D.‡

I am going to divide my discussion into four parts. I will just say a little about the history of rheumatic fever, going back to the first people who began to understand and describe this disease. Then I will talk a little bit about epidemiology, that is, how the disease is actually distributed, and the various factors of age, race, sex, and climate involved. Following that I wish to discuss the individual rheumatic child himself and to describe the clinical manifestations and treatment of rheumatic fever including what is being done on a national level in the care of rheumatic fever patients. Lastly, I will cover the subject closest to my own heart, and one that I think you all will be interested in, namely, the theories of the etiology of rheumatic fever.

HISTORY

I think it is always interesting and instructive in the discussion of any disease to try to reach back to the time when few thought about it or had any concept of its existence at all. Actually, in the writings of Hippocrates, there is a case report of a disease very much like what we call rheumatic fever today. This report demonstrates the close similarity of clinical manifestations regardless of the age in which an individual lives.

Hippocrates wrote[1]: (and you do remember he lived in 460 B.C.), "In those cases when there are pains and swellings about the joints, and they do not pass off in the manner of gout, you will find the bowels enlarged and white sediment in the urine, and if you inquire, the patient will admit that the temples are often pained, and he will say that he has nightly sweats. This disease forms in persons who in their youth had epistaxis and in whom it has ceased afterwards". Now that sounds a great deal, does it not, like polyarthritis in an adult who has grown up with a history of epistaxis in childhood and has had a recurrence of rheumatic fever? I do not have much doubt that it did exist in those days.

The next mention of the disease that is of interest was by Arateus[2] in 100 A.D. He describes rheumatism, and was apparently rather a

* From an informal talk presented to a group of Children's Hospital Residents and Howard University Medical Students at Howard University School of Medicine. Reproduced through the kind permission of Roland B. Scott, M.D., Professor of Pediatrics, Howard University School of Medicine.

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facetious person because he says, "Only the Gods understand it". Interestingly enough from the standpoint of our present concepts of etiology, the Greeks thought that rheumatism was due to an improper mixture of the humors of the blood. They believed that this improper mixture settled in the joints and caused the joint swelling and pain. This belief actually is not too far away from some of our current theories of etiology. You will find some references to polyarthritis in the writing of Aristotle, and Galen described arthritis very carefully and tried to account for its etiology.

But aside from these early writers and physicians, there is a great void in the discussion of the subject until we get down to the well known Sir John Sydenham[2], who made a life study of rheumatic fever. He not only described (in 1676), the chorea which bears his name but also described a juvenile form of polyarthritis. The term "rheumatic fever" was, of course, not yet used in those days. It was also Sydenham who coined the term, "scorbutical rheumation", which he used to describe the borderline cases which he could not classify definitely as gout or as an arthritis which left a lameness in the joints.

However, it remained for Hogarth, in the early eighteenth century to coin the term "rheumatic fever", and to pick the syndrome out of all the arthritides of the time. The cardiac symptoms were first described by Hogarth[2] in the late eighteenth century. However, he did not associate these symptoms with polyarthritis but felt that they were due to a secondary pneumonia. Pitcairn and Jenner[2], in the early nineteenth century were the first to associate rheumatic carditis definitely with rheumatic polyarthritis. An American physician by the name of Wells[3] first described rheumatic nodules in 1810. Bouillard[4] in 1836 wrote an excellent monograph on rheumatic fever (the disease is still known as Bouillard's disease in France today) in which he too recognized the association of cardiac manifestations with juvenile rheumatism.

The man who really gathered all these things together into a description of the clinical picture which can hardly be improved on even today was Cheadle[5] In 1889, he wrote an article in *Lancet* which described the association of tonsillitis, polyarthritis, carditis and chorea. He recognized erythema marginatum and other rashes which occur in the disease and, of course, also recognized the presence of rheumatic nodules. His article is quoted extensively in May Wilson's book[6] on rheumatic fever; and I suggest that those of you who are deeply interested in this disease read it.

It was a little later, in the early twentieth century, that the proponents of the streptococcus theory first put forward their ideas about the etiology of rheumatic fever. I have always felt that this was due largely to the influence of Pasteur; everybody was looking for a bacterial etiology of disease in those days. Be that as it may, the last fifty years have certainly been dominated by the idea that the streptococcus is associated with

rheumatic fever. The concept of focal infection was also introduced and is still with us. This theory postulates hidden niduses of infection which in some way cause "toxins" to circulate in the body and bring about the rheumatic state.

Aschoff, in 1904[7] described the Aschoff body and his original description is well worth reading if any of you are interested. Dr. Paul White quotes it *in toto* and some of his ideas and associations he made in describing the Aschoff nodule are most interesting. Within the past fifty years there are a few names that must be mentioned in discussing rheumatic fever. Coburn, of course, has done a great deal to further our knowledge of the streptococcal etiology. The shining exception to those who believe in the streptococcal theory is May Wilson[6] who has written a well known monograph on the subject of rheumatic fever. Homer Swift of the Rockefeller Institute believed in the streptococcal theory and did very good, fundamental work on the disease. Dr. T. D. Jones, I think, stands head and shoulders above them all as the person who did more for our understanding of rheumatic fever in the first part of this century. Dr. Ann Kuttner has done one of the finest pieces of work I can think of in proving the effectiveness of sulfonamide prophylaxis in preventing rheumatic recurrences. Others I will just mention include Massell, Ash, Headley and Paul, all of whom have done excellent work in the epidemiology of the disease. These are the outstanding names associated with rheumatic fever in our present century.

EPIDEMIOLOGY

Let us look at rheumatic fever as it stands on our planet today. Just what is it like, where is it and with what is it associated. It is a marvel to me that it took so long for a description of this disease to emerge, and yet I know that we, ourselves, are still missing many associations.

Be that as it may, in our own United States today, there are roughly a million people with rheumatic heart disease. Of these people, about 300,000 are school children and this figure is probably conservative. As you know, rheumatic heart disease is primarily a disease of the school age child. It does not occur very often below the age of four years, although it has certainly been described even in utero. At the end of the scale, first attacks have been described over the age of sixty. Generally speaking, however, this is a disease of the school-age child.

Based on personal examination of 8,000 school children, and 10,000 additional children examined by those who have worked with me, I would say that about 40 per cent of all children have what I have called "normal" murmurs, physiologic murmurs. At the very most, 2 per cent, (one per cent is probably a better figure) will have organic heart disease and of

these there will be four or five of rheumatic origin for each case of congenital origin. This is an important point to remember because so many children are erroneously diagnosed as having rheumatic heart disease on the basis of a rather loud systolic murmur. The vast majority of children with murmurs have no heart disease.

There is a difference in distribution of rheumatic heart disease in different sections of the country. Of course, in all sections of the country, there is no question about the fact that there is a familial incidence of rheumatic fever. Whether it is hereditary, as Dr. Wilson thinks (as a transmitted recessive gene), or whether it is due, as other people think to transmission from one member of a family to another, or a combination of the two, it is very difficult to say. Let us take, for example, New York, Texas and California. We know that the average age of death from rheumatic heart disease in New York is around 28. In Galveston, Texas the average age of death is around 42. Some very careful statistical analyses have revealed that California stands almost at the very bottom of the states in so far as the incidence of rheumatic heart disease in children is concerned; yet insofar as the incidence of rheumatic heart disease over the age of 20 is concerned, California runs in about the median sector of states.

There is no doubt then that the disease is more common in some areas and less common in others. In St. Francis Sanatorium, Long Island, rheumatic fever is seen in its most fulminating form, with subcutaneous nodules of months and years duration. Yet, in California, nodules are very uncommon. In Cincinnati I looked carefully for nodules for two years and found none at all. The point I want to make is that even though I say that subcutaneous nodules are more common in the New England states than in California, in the back of my mind there has always been a question (especially in view of May Wilson's work) whether it is just a matter of relatively slow development of the disease in climatically favored areas. This may explain the difference in age of death in New York and Texas, and the rapid change in incidence with increasing age in California. Perhaps the incidence is not so different but the severity and rate of development are different.

I have already mentioned the age factor. There is supposed to be a little more rheumatic fever in girls than in boys, but I can not help but feel that those figures are a little bit skewed. I have personally found equal distribution in girls and boys, although chorea is a bit more common in girls.

Rheumatic fever is prevalent in Europe and was considered to be so grave a health problem in England that the London County Council Rheumatism Scheme was organized a number of years ago to combat it.

I know very little about the oriental countries except that it is said that

the disease is not common in China. I have seen it in oriental children in this country. Rheumatic fever certainly occurs, and this is important, in areas where there is little scarlet fever. Dutch clinicians have reported what sounds like genuine rheumatic fever on some of their island possessions where scarlet fever is practically unknown.

Another curious and important thing is the tendency for rheumatic fever to recur. Following the original episode of rheumatic fever, the chances of recurrence are probably five times as great as they were before the original attack. Our whole approach to the control of the disease is based on this point, of course.

CLINICAL MANIFESTATIONS

Let us now look at how rheumatic fever may appear and act in the individual person. This is one disease that one must live with to understand. In my earlier days in Cincinnati, I thought I knew something about the disease, but the more deeply I have gone into it, the more cautious I have become and the humbler I am about making positive statements about it.

Most investigators (Dr. Wilson is the most prominent exception) feel that rheumatic fever is closely allied to streptococcal disease. Certainly, there is no doubt that the so-called "classical" picture of phase one, phase two, and phase three (streptococcal disease; latent period of two to six weeks; active rheumatic fever) does occur. My own experience leads me to believe that rheumatic fever so frequently occurs in other ways that we can hardly call this the "classical" picture. When it does occur in this way it is important to remember that there is a latent period between the streptococcal infection and the onset of rheumatic manifestations; there are many symptoms associated with acute streptococcal disease which may be confused with rheumatic fever but which certainly are not rheumatic fever. The arthralgias accompanying streptococcal infection are the prime example.

In describing the disease, I can think of no better way to present it to you than to mention patients I have seen myself. One was a little five year old girl in Colorado, who was admitted to the hospital with one joint swollen. The temperature was elevated and it looked like rheumatic fever even though she had no evidence of carditis. We put her to bed and observed her for 24 to 48 hours before doing anything. This is a point of crucial importance in observing and managing polyarthritis in children when it does occur. Put them to bed and observe for the so-called typical migratory polyarthritis before you jump in with any kind of treatment at all because if you once make a diagnosis of rheumatic fever, you are committed to a lifelong regime. In any event, this child had a classical migra-

tory polyarthritis, responded beautifully to salicylates, and recovered with no heart damage at all. That was ten years ago.

A surprising number of cases are like that: a mild attack of polyarthritis, no evidence of carditis and complete recovery. They are, of course, given antibiotics prophylactically and may have no further difficulty.

At the opposite extreme is a little five year old boy I recently observed, who was admitted with full-blown, active carditis, in congestive failure, and who in spite of all measures died in about six weeks.

In between these two, the mild and the fulminating form, there are a number of different clinical pictures. Some have chorea as a presenting manifestation. Initially there may be nothing more than emotional instability, a very common early complaint in chorea. Of course, with adequate sedation, the disease usually subsides spontaneously with about six weeks of bed rest. As you know, chorea is not so often complicated with heart disease as are the other manifestations of rheumatic fever. I do feel, however, that chorea is definitely a rheumatic manifestation. There are others who do not, but I do not think that this is in line with most of the evidence.

In thinking of the many curious ways in which I have seen rheumatic fever develop, I remember a young man coming down from the mountains in Colorado and telling me that for a week or two he had been feeling sleepy and having some pain in his right side. It sounded more like encephalitis than anything else: no history of streptococcal infection; nothing that even remotely resembled polyarthritis or chorea; just a history of drowsiness and pain in the right side. Of course, it developed under clinical examination that he was in acute congestive failure with a large tender liver and had very definite active rheumatic carditis.

Another young man who had had an initial attack of rheumatic fever was working, of all things, loading freight cars. One day he sat down in the open doorway of a freight car to cool off. The next day he had polyarthritis and was admitted with a full-blown recurrence of rheumatic fever with severe carditis and heart failure. The association of recurrences with sudden chilling is an interesting thing. It has been noted by people who have studied the disease from the very beginning. The clinicians back in the nineteenth century noticed that these attacks of polyarthritis did have something to do with chilling.

Another child who we saw walked into our clinic with full-blown rheumatic myocarditis and pericarditis and not only manifestations of mitral and aortic disease, but tricuspid disease as well. He was hospitalized and died within 48 hours, and yet he was ambulatory before we saw him.

Insidious carditis may occur: a little girl who had no findings, not even an elevated sedimentation rate, no fever, and a little tachycardia. I watched

her for a year feeling she had carditis even though she had none of the laboratory findings usually associated with rheumatic fever. She eventually had aortic insufficiency. This child serves to illustrate my original statement that rheumatic fever is not a "classical" disease but may develop in a great many different ways.

Erythema marginata, I am sure you have heard, is more or less a minor manifestation. In eliciting a history of erythema marginata in any child who has rheumatic fever, I make a practice of asking the parent if the child has had a rash that looked like ringworm, for that is exactly what erythema marginata looks like. It is a curious eruption. It is frequently present in the morning and absent in the afternoon, and when observed is usually present on the flexor surface. It is considered to be one of the rather benign manifestations of rheumatic fever, and is thought to indicate a good prognosis. I might also say that erythema marginata can occur without any other evidence of rheumatic fever. This has been described and I have personally seen two cases.

Rheumatic nodules are another thing entirely. I do not believe I have ever seen them, except once, in the absence of carditis. They are rather hard and initially look like little pieces of rice under the skin, although they may attain the size of a small marble. They are seen a lot better than they can be felt and most commonly occur over the triceps tendons although they may occur on any of the tendons. They are usually painless. They may occur on the scalp where they are bigger, softer and may be painful. Children do not like to lie on their heads when they have rheumatic nodules on their scalp. They are, as I have said, usually associated with severe fulminating rheumatic carditis.

In examining the heart of a patient one suspects of having rheumatic fever, the first thing one needs is the proper type of stethoscope (Rieger-Bowles). This is not necessarily true in diagnosing congenital malformations of the heart but is certainly true for rheumatic heart disease where auscultation is an all-important procedure for making the diagnosis. I believe it is the trained ear, more than anything else, that can distinguish between the presence and absence of carditis in a child. The first thing to listen for is a diminution in the first heart sound. It becomes mushy and impure in character. At the same time, the second sound becomes somewhat accentuated. The third heart sound if previously present, becomes a little bit accentuated, or if not previously present, appears.

Tachycardia may or may not be present. I have seen some of the most severe kinds of rheumatic carditis without tachycardia. Personally I have never seen a child with active rheumatic carditis who had sinus arrhythmia. For some reason or another, they just do not occur together. Premature systoles do occur rather frequently. They are more likely to be nodal in type.

In early rheumatic carditis, one unusual type of systolic murmur may occur. It is not described in textbooks; only students of rheumatic fever are familiar with it. It occurs toward the left sternal border and sounds more like a murmur of congenital heart disease, and very much like the murmur that occurs in complete heart block. I have never attempted to correlate this murmur with the presence of first degree A-V block but that is a point I have in the back of my mind to watch.

These children also often get a certain look when they have active carditis. You have to see it to appreciate it. They look very sick even though their charts do not indicate it. Later on they may develop more obvious evidence of active carditis, the rather soft, very typical murmur of mitral insufficiency.

We might dwell a minute on the murmur of mitral insufficiency. Whatever you may read to the contrary it can quite definitely be distinguished from other systolic murmurs in almost every case. A good way to reproduce this murmur is to place the bell of the stethoscope over the back of your hand and run your finger along the adjacent surface and listen. You will hear a very excellent reproduction of the murmur of mitral insufficiency.

The murmur of mitral insufficiency varies a great deal in the child with active rheumatic heart disease but in its early form it is still quite characteristic, and is heard best at the apex with transmission to the axilla and usually rather well to the back. It is not transmitted to the neck but is usually increased in intensity after exercise. This latter point is true of almost all murmurs, however. Exercise is not helpful in differentiating systolic murmurs, but only in amplifying them.

The heart disease may progress a little further and the auscultatory findings of mitral stenosis may develop. Initially, of course, there may be a mid-diastolic murmur which means nothing at all as far as organic changes are concerned. Very commonly, in the presence of an initial attack of active rheumatic carditis, a frank, loud, mid-diastolic rumble will develop. The intensity of this rumble may actually increase after a few days but of course does not mean mitral stenosis at all.

Pericarditis is much more common than is generally appreciated. Listen carefully for it. It usually is represented by a little scratchy to and fro sound which is completely distinct from the murmurs I have previously described. It may be present for a few days and then may disappear entirely.

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